

A 2-year-old boy is brought by his mother to the emergency department because of a high-grade fever which "does not go away" with acetaminophen. For the last four days, the child has been very irritable and is crying a lot. He is also pulling his ear and not eating well. He has been generally well, other than the occasional sore throat this season. His temperature is 38.8° C (102.2° F), blood pressure is 90/60 mm Hg, pulse is 119/min, and respirations are 24/min. He appears well nourished, but is irritable. Physical examination reveals enlarged cervical lymph nodes and splenomegaly. The tympanic membranes are inflamed. CBC shows:

WBC	81,100 mm ³
Hemoglobin	8.0 g/dL
Hematocrit	25%
Platelets	16,000 mm ³
Blast forms	80%
Prolymphocytes	10%
Lymphocytes	10%

The blast cells have condensed nuclear chromatin, small nucleoli and scant agranular cytoplasm. Subsequent histochemical staining reveals strongly positive periodic acid Schiff (PAS) reaction. No Auer rods were seen. Which of the following is the most likely diagnosis?

- ☐ A. Burkitt lymphoma
- ☐ B. Acute myelocytic leukemia
- ☐ C. Prolymphocytic leukemia
- ☐ D. Acute lymphoblastic leukemia
- ☐ E. Myelodysplastic syndrome

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- ☐ A. Burkitt lymphoma [6%]
- ☐ B. Acute myelocytic leukemia [9%]
- ☐ C. Prolymphocytic leukemia [3%]
- ☒ D. Acute lymphoblastic leukemia [79%]
- ☐ E. Myelodysplastic syndrome [3%]

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✓ ☒ D. Acute lymphoblastic leukemia [79%]

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Explanation:

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Acute lymphoblastic leukemia (ALL) is the predominant type of leukemia in children from ages 2-10 years. Approximately 30-50% of patients present with infections, and about half present with lymphadenopathy and splenomegaly. Lymphoblasts are typically seen on the peripheral smear. Varying degrees of anemia, neutropenia, and thrombocytopenia have been noted. The presence of more than 25% lymphoblasts in the bone marrow is diagnostic. Lymphoblasts lack peroxidase positive granules but often contain cytoplasmic aggregates of periodic acid Schiff (PAS) positive material. Immunostaining for terminal deoxynucleotidyltransferase (TdT) is positive in more than 95% of patients. TdT is expressed only by pre B and pre T lymphoblasts.

Myeloblasts on the other hand contain peroxidase positive material.

(Choice A) Burkitt lymphoma is a neoplasm of mature B cells. It is associated with the Epstein-Barr virus infection. Most patients present with either a mass involving the mandible or abdominal viscera. High mitotic index is typical. Histological examination shows characteristic "starry sky appearance". It is a very aggressive tumor but responds well to the high dose chemotherapy.

(Choice B) In acute myelocytic leukemia (AML), the predominant cells are of myeloid origin. Auer rods are specific for AML.

(Choice C) Prolymphocytic leukemia is a CLL variant. It is characterized by massive splenomegaly, high lymphocytosis, and lymphadenopathy.